Aldurazyme® (laronidase) (Intravenous)

Effective Date: 02/01/2020
Review Date: 01/15/2020, 1/28/2021
Revision Date: 01/15/2020, 1/28/2021
Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

I. **Length of Authorization**

Coverage will be provided for 12 months and may be renewed.

II. **Dosing Limits**

A. **Quantity Limit (max daily dose) [NDC unit]:**
   - Aldurazyme 2.9 mg vial: 92 vials every 28 days

B. **Max Units (per dose and over time) [HCPCS Unit]:**
   - 667 billable units every 7 days

III. **Initial Approval Criteria**

Coverage is provided in the following conditions:

MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

- Patient is 6 months of age or older: **AND**
- Patient has absence of severe cognitive impairment: **AND**
- Documented baseline value for urinary glycosaminoglycan (uGAG) has been obtained: **AND**
- Documented baseline values for one or more of the following have been obtained:
  - Patients 6 years or greater: percent predicted forced vital capacity (FVC), 6-minute walk test, joint range of motion, left ventricular hypertrophy, growth, quality of life (CHAQ/HAQ/MPS HAQ): **OR**
Patients 6 months to less than 6 years: cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC, and/or 6-minute walk test; AND

Mucopolysaccharidosis I (MPS I) †

- Patient has a definitive diagnosis of MPS I confirmed by one of the following:
  - Detection of biallelic pathogenic mutations in the IDUA gene by molecular genetic testing; OR
  - Detection of deficient activity of the lysosomal enzyme α-L-iduronidase (IDUA); AND
- Patient has one of the following diagnoses:
  - Hurler (severe) or Hurler-Scheie (attenuated) forms of disease; OR
  - Scheie (attenuated) form of disease with moderate to severe symptoms

† FDA approved indication(s) Φ Orphan Drug

IV. Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet indication-specific relevant criteria identified in section III; AND
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis and severe hypersensitivity reactions, acute respiratory complications, acute cardiorespiratory failure, severe infusion reactions, etc.; AND
- Patient does not have progressive/irreversible severe cognitive impairment; AND
- Patient has a documented reduction in uGAG levels compared to pretreatment baseline; AND
- Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following:
  - Patients 6 years or greater: stability or improvement in percent predicted FVC and/or 6-minute walk test, increased joint range of motion, decreased left ventricular hypertrophy, improved growth, improved quality of life (clinically meaningful change in the CHAQ/HAQ/MPS HAQ disability index); OR
  - Patients 6 months to less than 6 years: stability or improvement in cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC and/or 6-minute walk test
V. **Dosage/Administration**\(^1,2,5,6\)

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tbody>
<tr>
<td>Mucopolysaccharidosis I (MPS I)</td>
<td>0.58 mg/kg of body weight administered once weekly as an intravenous infusion</td>
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</table>

VI. **Billing Code/Availability Information**

**HCPCS Code:**

J1931 – Injection, laronidase, 0.1 mg; 1 billable unit = 0.1 mg

**NDC:**

Aldurazyme 2.9 mg/5 mL single-dose vial: 58468-0070-xx

VII. **References**


**Appendix 1 – Covered Diagnosis Codes**
<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
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<tbody>
<tr>
<td>E76.01</td>
<td>Hurler’s syndrome</td>
</tr>
<tr>
<td>E76.02</td>
<td>Hurler-Scheie syndrome</td>
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<tr>
<td>E76.03</td>
<td>Scheie’s syndrome</td>
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**Appendix 2 – Centers for Medicare and Medicaid Services (CMS)**

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Articles may exist and compliance with these policies is required where applicable. They can be found at: [http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx](http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx). Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/Article): N/A

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<td>5</td>
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